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ASSOCIATED WITH PROGRES-
SIVE MUSCULAR ATROPHY.
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TWO CASES OF NERVOUS DISEASE: (1) A CASE OF PERIPHERAL NEURITIS WITH SYMPTOMS OF BULBAR PARALYSIS. (2) A CASE OF LOCOMOTOR ATAXY ASSOCIATED WITH PROGRESSIVE MUSCULAR ATROPHY.

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(1) A CASE OF PERIPHERAL NEURITIS WITH SYMPTOMS OF BULBAR PARALYSIS.

G. M., aet. 29, a farmer, was admitted to Ward 7 of the Royal Infirmary, on January 14th, and he died on January 21st, 1896.

The commencement of the illness seems to have dated back to January, 1895, when the patient had a sore throat lasting for several days. It was not at the time thought to be diphtheritic, and there was no diphtheria known to be in the neighbourhood. But some weeks later the patient found that he had difficulty in masticating his food, and he was quite sure that this was due to loss of power in the muscles of the jaw. In the month of March the soft palate became involved, giving rise to a nasal quality in the speech and allowing fluids to regurgitate through the nose. The eyelids were likewise affected, for they could neither be firmly closed nor completely raised, and the patient had the appearance of a slight double ptosis. These symptoms were more marked in the right than in the left eye. At this time he also complained of his legs and arms being "weak." He said he was easily tired, that his legs would give way under him and that he several times fell.

This condition of the limbs, however, improved with electrical treatment and he was ultimately able to walk quite well.

For some months prior to admission there was difficulty in swallowing, which seemed to gradually get worse. More recently loss of power in the lips and tongue developed, and there was difficulty in chewing and controlling the food while in the mouth. At the time of admission to the ward the tongue, though it presented no apparent atrophy, could not be protruded to any extent and its movements within the mouth were very limited. Neither could the lips be closed even by a strong effort, and saliva was always dribbling from out the corners of the mouth. The patient could make no whistling sound and when he attempted to do so the air escaped freely from between the lips. The lower jaw seemed to be too loosely articulated, it tended to fall forwards, and the patient seemed to find it impossible to keep the upper and lower dental arches in apposition. At this time he could still swallow, though with difficulty, and his speech, while indistinct, was quite intelligible. But during the week subsequent to admission the symptoms rapidly got worse, so that by the 18th inst., he had to be fed by the stomach tube and his articulation was so much affected that he could only be understood when he conveyed his wishes in writing. The respiratory movements by this time were mostly diaphragmatic, although on admission to the ward there was still considerable intercostal action. Finally the breathing became entirely abdominal and death seemed to supervene from failure of respiration.

While in hospital there was little loss of power to be noted in the muscles of the arms and legs. The grasp in both hands was good, and the patient could stand with his feet together and the eyes shut without any difficulty. He could walk quite well, and while the gait was slow and deliberate, it presented no characteristic abnormality. The tendon reflexes were normal and there was no ankle clonus. The plantar and cremasteric reflexes were not obtained, but the abdominal reflex was active. There was no reaction of degeneration to be found in the muscles of the face, arms, shoulders or legs. There was no impairment of general sensibility and the special senses seemed quite normal. The movements both of the internal

and external muscles of the eyeball remained normal. The patient complained of "weakness" in his back and he seemed to have difficulty in raising himself from the dorsal decubitus without first turning on to his side. One could not, however, be certain that this symptom was due to actual paralysis in any of the muscles and the trunk.

The heart and lungs showed no signs of disease, but the urine always contained a certain amount of albumen.

At the *post-mortem* examination the macroscopic appearances were found to be perfectly normal, except that both kidneys were seen to be in the early stage of interstitial nephritis. The cord, medulla, pons, and the ixth, xth, and xiith nerves were examined microscopically by Weigert's and Nissl's methods. The right and left vagus and right and left glosso-pharyngeal nerves showed a fairly well-marked degeneration, with great increase of the internodal nuclei. Both right and left hypoglossal, however, seemed quite normal. Weigert's staining showed no degeneration in the motor tracts of the cord, medulla, or pons; and the ganglion cells in the anterior horn of the cord, in the hypoglossal nucleus, the nucleus ambiguus, and in the facial nucleus seemed little altered. These cells throughout were well shaped, they had central nucleus, and there was no apparent absence of processes. The Nissl bodies, however, were not so well defined as might be desired, but there was certainly no advanced chromatolysis to be seen; and if there was any fragmentation of the Nissl granules, it was very slight. The defect in the definition of these Nissl granules was probably due to some slight error in fixing or in the stain itself, but examination of a large number of sections of the cord and medulla convinces me that the ganglion cells of the anterior horns of the xiith nucleus were practically normal. One cannot speak with the same assurance of the cells of facial nucleus and the nucleus ambiguus. There were not the same number of sections at these levels to examine, and it is difficult from a few transverse sections (especially in the case of the xth nucleus) to give a definite opinion as to the condition of the whole nucleus. We may take it, however, that the cells of the viith and xth nuclei showed little, if any, change.

Microscopic examination of the muscles of the pharynx and tongue showed them to be quite normal.

Viewing this case, then, in the light of the *post-mortem* examination, it seems, with little doubt, to be one of peripheral neuritis, probably of diphtheritic origin. The microscopic appearances explain the clinical signs and symptoms, except that it is difficult to understand the absence of degeneration in both hypoglossal nerves. The presence of at least a certain amount of paresis in the tongue seemed undoubted during life; but, on the other hand, it is to be noted that careful microscopic examination of that muscle showed no corresponding degeneration in its fibres, such as one would look for in a case of peripheral neuritis. Possibly, as is so often the case in diphtheritic paralysis, only some of the fibres in the nerve were affected, and these towards their distal terminations, where degeneration most often shows itself. It is, therefore, much to be regretted that both hypoglossals were not examined through a greater extent of their distribution, and stained also by Marchi's method. It is quite certain, however, that in the portions of the nerves examined there was no increase of the internodal nuclei, and that the axis cylinders were quite intact. These points were well shown in the sections stained with methylene blue and Congo red.

If this case, then, is one of peripheral neuritis, its special interest would seem to lie in the fact that, with histological changes confined to the peripheral nerves, we have a grouping of symptoms almost identical with those found in bulbar paralysis. As in that disease, the onset was gradual. The lips, tongue, soft palate, pharynx, and larynx were all involved. It is true that the lips and tongue were affected late in the disease, and that there was no atrophy of the tongue. But it sometimes also happens this way in bulbar paralysis. In bulbar paralysis, too, we sometimes have the muscles of the upper part of the face and of the jaw affected just as in this case. The symptoms, therefore, closely resemble those found in bulbar paralysis; and the above record may be of value in that it might suggest an explanation of these rather obscure cases of "chronic bulbar paralysis without anatomical lesion."

(2) A CASE OF LOCOMOTOR ATAXY ASSOCIATED WITH PROGRESSIVE MUSCULAR ATROPHY.

J. O'B., aet. 48, a labourer, was admitted to Ward 7 of the Royal Infirmary, on June 19th, 1896.

The notes of this case are rather scanty as the patient died within twenty-four hours of admission. He had been admitted a month before to a surgical ward on account of a bruise on the right hip, and he was transferred to the medical house as a case of progressive muscular atrophy. During his residence in the surgical wards his symptoms seem at no time to have suggested a diagnosis of locomotor ataxy.

The history the patient gave was that for ten years past he had been aware of atrophy in his hands, which had gradually been getting worse. On examination both hands presented the typical "bird-claw" appearance. The thenar and hypothenar eminences were quite gone and there was much atrophy in the interossei muscles. The upper-arm muscles were also involved to a certain extent. There was considerable loss of power in both hands and arms, and tremor in the hands was present. The tactile sense was much impaired in the parts of the hands supplied by the ulnar nerve.

The muscular power of the legs was good, but the tactile sense was lost in both feet and much diminished in the legs as high up as the knees. The plantar and knee reflexes were gone, but the cremasteric and abdominal ones quite active.

The pupils were unequal and neither contracted to light. The sense of smell was somewhat deficient, but the senses of sight, taste, and hearing were quite normal.

For a period of four years patient voided his urine in bed, but he said he could retain it during the day. He had been a heavy drinker for many years, and there was trembling of the lips and tongue, especially when he was speaking. The memory, also, was somewhat impaired.

On May 23rd delirium tremens developed and lasted for five days, in spite of treatment by large doses of chloral and bromide of potassium. This illness left him very feeble.

Shortly after admission to the medical wards convulsive

seizures supervened. These were clonic in character, and generalised, but they affected the extensor more than the flexor muscles of the body. The duration of each convulsion varied much, the longest being three hours and the shortest about one minute. These seizures continued, on and off, for about twenty-four hours, at the end of which time the patient died.

The *post-mortem* examination revealed infarctions in the left kidney and a condition of interstitial nephritis in the right. There was some excess of fluid in the soft membranes of the brain, and some ecchymosis on the right frontal lobe and over the left motor area on both sides of the fissure of Rolando. Otherwise the macroscopic appearances were practically negative.

Microscopic examination.—The nerves of both arms were stained with osmic acid and with carmine. They all showed marked degenerative changes, with great increase of the inter-nodal nuclei and connective tissue elements, some of the nerves being just like strands of fibrous tissue.

The cord, medulla and pons were stained according to the methods of Weigert and Nissl. With the *Weigert* stain there was seen to be a well-marked degeneration in the posterior columns of the cord. In the lumbar region this involved both the postero-external and the postero-internal columns; but passing upwards it gradually confined itself to the postero-internal till in the cervical region it was only this tract that was affected. The posterior roots in the lower cord were much sclerosed but in the cervical region they were apparently unaffected. The anterior roots in the cervical region seemed practically normal. There was no apparent degeneration in any of the antero-lateral tracts, neither did the motor fibres in the medulla or pons show any abnormality.

In the specimens stained by Nissl's method special attention was paid to the appearances of the motor ganglion cells. Sections were examined from six different levels of the cervical cord, four of these being from the cervical enlargement, and the most striking feature of these sections was the small number of cells showing any marked pathological change, the vast majority being well shaped, with central nucleus, and their

processes intact. Many of them contained a considerable amount of yellow pigment, but in only a very small proportion was anything like a chromatolytic change to be seen. A count of the cells in the anterior horns was made, and although the number in all the sections through the cervical enlargement was below that of the normal cord, still the number of cells present was much greater than one would look for with such an extreme degree of muscular atrophy as was present in this case. The mesial group of cells was not considered in the count, and the antero-lateral and postero-lateral groups were as a matter of convenience taken together as one. The average number of cells in this lateral group throughout the cervical enlargement was 36·6 as against 54, the average number found in a healthy young adult. But it is to be here noted that there was also a diminution in the number of cells in the anterior horns in the lumbar region (30 as compared with the normal 47), though there was no apparent corresponding atrophy of the muscle-fibres supplied by these cells. I take it, then, that in estimating the number of ganglion cells in such a case as this—a man, aged 48, and who, from our point of view, was really much older—we must not compare his cord with that of a young adult, but rather with that of a much older man and one who has not recently been living an active life; and I find in counting the cells in other cases that the standard of the young adult is too high for most of the cases over 50 years that one examines.

I take it, then, that in the cervical enlargement there was no atrophy of ganglion cells corresponding to the very great atrophy in the muscles of the hands and arms, or to the degree of degeneration found in the nerves supplying these muscles. The absence, too, of degeneration in the antero-lateral tracts of the cord, and in the corresponding tracts of the medulla and pons corroborates this view. And so the conclusion that is forced upon us is that the primary lesion has been one of peripheral nerves. But this corresponds with the researches of Dégérine which point to the muscular atrophies which sometimes complicate locomotor ataxy, as being usually due to degeneration in peripheral nerves, though it is opposed to

the view of Gowers, who tells us that any extensive muscular atrophy found in that disease "is probably due, as a rule, to degeneration in the anterior cornua."

From this point of view, then, this case of ours has presumably been primarily one of locomotor ataxy, and the progressive muscular atrophy a complication. But on the other hand, it would be impossible to insist that it was not the progressive muscular atrophy that was the primary disease, for the atrophy was always the most outstanding feature of the case, and it had been present for at least ten years. It is quite conceivable, however, that locomotor ataxy may have been present for a longer period still, and yet not have obtruded itself specially on the notice of a patient whose habits of life would not lead to accuracy of observation, and whose mind for so many years was fixed on the atrophy and loss of power in his arms and hands.

